Infected aneurysm of sinus of Valsalva Report of a case with involvement of all three sinuses

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A case of infected aneurysms of all the three sinuses of Valsalva is described, and the clinical course and possible morphogenesis of this rare disorder are discussed

Aneurysm of the sinus of Valsalva is not an uncommon disorder. Its incidence has been estimated at 3.5 per cent of surgical congenital heart diseases, and the condition can be diagnosed with a fair degree of certainty on clinical and haemodynamic grounds and is amenable to surgery in a fair number of cases (Taguchi et al., 1969). The majority of the cases show involvement of one, and some cases show involvement of two sinuses, while cases with aneurysms of all three sinuses are distinctly rare, and to date only five cases have been reported.

One such case, with aneurysms of all the three sinuses, is reported here.

Case report

A 28-year-old man was admitted in September 1968 with a history of severe dyspnoea which was first noticed during coitus eight weeks earlier and again a week later. Since then it had persisted. There was no chest pain or fainting or giddiness. There was a past history of a penial sore 5 years ago, and serological tests for syphilis had been reported positive at another hospital. He was referred to this Institute as a case of rheumatic aortic incompetence with congestive cardiac failure. On examination he had a collapsing pulse, and a blood pressure of 130/50 mm. Hg. Apex beat was in the 5th intercostal space 2.5 cm. outside the midclavicular line. Both heart sounds were normal, and an ejection click was audible at the apex and the left second space. There were mid-systolic and early diastolic murmurs at the aortic area. The mitral area revealed pansystolic and mid-diastolic murmurs and a third sound. The patient had slight clubbing, the spleen was not palpable, there was no fever, and other systems showed signs of congestive cardiac failure, mainly on the left side. The electrocardiogram showed complete left bundle-branch block, occasional ectopic beats, and a prolonged PR

interval. Chest x-ray showed enlarged heart and evidence of pulmonary congestion. Five consecutive blood cultures were sterile, and the sixth one grew Staph. pyogenes and Mima polymorpha. Urine was repeatedly negative for red blood cells, and a diagnosis of bacterial endocarditis could not be established. After the control of cardiac failure a haemodynamic study was performed. It showed a moderate increase in left ventricular enddiastolic pressure, absence of any gradient across the aortic valve, and moderate pulmonary arterial hypertension. The patient was advised to have aortic valve surgery but refused, and he was allowed to go home after a month in hospital on digoxin and diuretics, after an antiluetic course of procaine penicillin 400,000 units intramuscularly twice daily for a fortnight. He stopped all medication at home and was readmitted 3 weeks later in a state of severe congestive failure. There was no significant change in the local cardiac signs, and in spite of medical treatment he died 3 days later. The entire course of his illness thus lasted about 6 months.

Necropsy The heart weighed 430 g. and it showed left ventricular hypertrophy and dilatation. The aortic valve showed abnormally deep sinus pockets - up to 3.5 cm. in depth. The left coronary sinus appeared as a bag of laminated thrombus extending downwards and encroaching upon the aortic leaflet of the mitral valve. The noncoronary sinus extended down slightly to the left of the membranous trigone, while the right coronary sinus overrode the left ventricular outflow tract downwards and projected as a bulge into the right ventricle. The free rims of all the three cusps were thickened, rolled up, and showed a few nodular, smooth, and firm excrescences. Of the original aortic leaflets only the commissures could be recognized separately; the surface distinction between the cusps was obscured by smooth scar tissue. This scarring extended down into the outflow tract of the left ventricle (Fig. 1, 2, and 3).

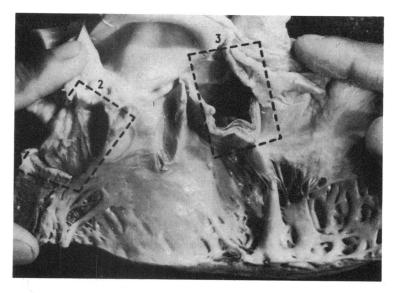
FIG. I Gross picture of left ventricle to show the aneurysmal pouches projecting down. Fig. 2 and 3 are from blocks taken from the outlined areas 2 and 3.

Microscopy confirmed the healed and healing endocarditis, the elastica of the aortic valve could be made out in the scar tissue which formed the anterior wall of the aneurysms. No bacteria were seen in the vegetations. Above the ring, the aorta was normal apart from a few fatty streaks. The mitral valve was normal except for mild fibrotic thickening of the aortic (anterior) leaflet. The right heart was normal. Moderate passive congestion was seen in the liver and lungs, which did not show pulmonary arteriosclerosis.

Comment

Till the early 1960's it was customary to distinguish between 'congenital' and 'acquired' aneurysms of the sinus of Valsalva. This practice has since ceased, for, as Hudson (1965) points out, it is virtually impossible in most cases with infection to rule out a congenital basis, it might therefore be better to refer to 'infected and uninfected' aneurysms. However, in a recent report, Taguchi et al. (1969) have maintained the distinction between congenital and acquired aneurysms of the sinus of Valsalva. Presenting their experience of surgical correction of this disorder, in 45 cases of congenital aneurysms, these authors have proposed a detailed anatomical classification of the aneurysms into 6 main and 16 subdivisions. The congenital ones, according to Eliot, Wolbrink, and Edwards (1963) result from a congenital weakness at the attachment of the aorta to the annulus fibrosus, so that the tissues here give way. Syphilis has occasionally been mentioned as a cause of these aneurysms (Ostrum et al., 1938; Micks, 1940), but most authorities do not accept this view. Acute bacterial endocarditis resulting in aneurysm of the sinus of Valsalva is well known (Björk and Björk, 1965).

As has already been mentioned, only 5 cases have been reported so far in which all the three sinuses of Valsalva showed aneurysms. In the report of 45 cases from Japan by Taguchi et al., referred to above, all cases had single sinus aneurysm. Micks in 1940 reviewed 3 such cases reported previously (those of Carpentieri from Naples, 1912, Barnscheidt from Bonn, 1920, and Habán from Budapest, 1937) and added one case of his own. The fifth patient was reported by Basabe, Hojman, and Rosemblit (1954). The case being reported here almost faithfully repeats the clinical features of the previous 5 cases. Symptom-free young adults in rapidly



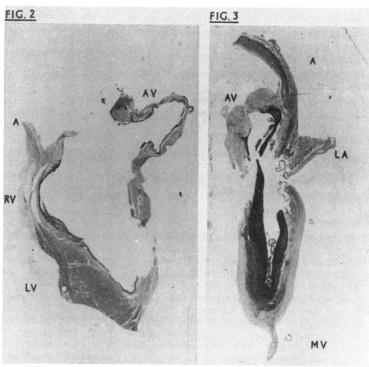


FIG. 2 Low magnification photomicrograph of the left coronary sinus and valve cusp. The downward bulge of the aneurysm over the ventricular muscle can be seen. (A: aorta, AV: aortic valve, RV: right ventricle, LV: left ventricle).

FIG. 3 Low magnification photomicrograph of the right coronary sinus and cusp. The nodular vegetations at the free edge and the laminated thrombus in the cavity are seen. (LA: left atrium, MV: mitral valve, anterior leaflet).

progressive aortic failure, which ends in death within a few months or weeks, appear to be the usual pattern of this disorder. The diagnosis has always been established at necropsy and, unlike the aneurysm of the single sinus, surgical correction does not appear feasible

From the histological study, it seemed that the infective process had eroded the crypts of the cusps and the adjoining walls of the sinuses without causing perforation of the cusp substance into the ventricle. The presence of the elastic lamina of the cusp in the upper portion of the anterior aneurysmal walls supports this. Though there was a history suggestive of syphilitic infection 5 years earlier, the morphological examination of the aorta failed to show any evidence of syphilitic process. It is likely that this patient had an infective endocarditis which was controlled by the penicillin given for treatment of syphilis. The bacterial infection also presumably brought about the heart failure.

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